SHORT REPORTS

Pregnancy in women with Friedreich's ataxia

Friedreich's ataxia is an uncommon autosomal recessive disease of the nervous system affecting one in 50 000 people. In many cases its onset occurs in adolescence. As well as having severely disabling neurological symptoms 8-18% of patients have diabetes mellitus and 60-90% have electrocardiographic abnormalities, of whom one third have cardiac symptoms and signs.^{1,2} Despite these severe complications affecting several organ systems no study of the effects of Friedreich's ataxia on pregnancy and its outcome has been reported previously.

Patients, methods, and results

I sent a questionnaire to women of childbearing age with Friedreich's ataxia on a list obtained from the Friedreich's Ataxia Association. A total of 18 women who had been pregnant were identified from this list, of whom three were excluded because the disease had been diagnosed after their pregnancies. In addition I myself investigated two cases. Of the 17 patients, three had identifiable cardiac signs: two had hypertrophic obstructive cardiomyopathy and one recurrent tachycardia and electrocardiographic abnormalities. One patient had insulin dependent diabetes mellitus, and one had yielded an abnormal result to a glucose tolerance test, although she was not taking any specific drugs.

The table shows the obstetric outcome of the 17 patients. There were no perinatal deaths. All women had been delivered of live babies in excess of 36

Details of pregnancy in 17 patients with Friedreich's ataxia

Case No	Age at diagnosis of Friedreich's ataxia (years)	Parity	Delivery (all >36 weeks)			Associated conditions
			Spontaneous	Forceps	Epidural	
1	12	1+0	1	—	1	Hypertrophic obstructive cardiomyopathy
2	7	1+0	1	—	—	Insulin dependent diabetes
3	21	1+0	1	—	—	Supraventricular tachycardia
4	17	1+0	1	—	—	
5	23	2+0	2	—	—	
6	5	1+0	1	—	—	Hypertension induced by pregnancy
7	18	2+0	1	—	1	Hypertension induced by pregnancy
8	16	3+0	3	—	—	
9	20	1+0	—	1	—	Hypertrophic obstructive cardiomyopathy
10	15	2+0	2	—	—	Abnormal glucose tolerance test; gestational diabetes
11	15	1+0	1	—	—	
12	18	1+0	1	—	—	
13	18	2+0	2	—	—	
14	25	2+0	2	—	—	
15	16	2+0	2	—	—	
16	19	2+0	1	—	—	
17	29	1+1*	1	—	—	

*Termination of pregnancy and sterilisation at five months.

weeks' gestation. Five women had been offered termination of pregnancy because of their condition and four had refused; all four were delivered of a live baby with no obstetric complications. Two patients had hypertension induced by pregnancy but subsequent normal deliveries. Three women received epidural anaesthesia: two, one of whom had hypertrophic obstructive cardiomyopathy, had a normal delivery and one had a forceps delivery. One of the 16 women without diabetes mellitus developed gestational diabetes during her pregnancy.

Contraceptive follow up for the 17 women varied. Two were sterilised, having completed their families; six used a combined oral contraceptive pill; three used an intrauterine contraceptive device; and the remainder used barrier methods of contraception.

Comment

Although Friedreich's ataxia is a serious disease with serious cardiovascular and metabolic problems, the reproductive performance of the 17 women was good. They did not seem to have been prone to antenatal complications, especially premature labour and hypertension induced by pregnancy. Thus a normal delivery should be expected in women with Friedreich's ataxia; when epidural anaesthesia is required it should not be withheld purely because of the condition. Friedreich's ataxia itself is not an indication for termination of pregnancy. Cardiovascular examination is

mandatory in all patients, and early referral to a cardiologist is recommended. In those who have heart disease electrocardiographic monitoring throughout labour should be recommended.

Couples of whom one or both are affected with Friedreich's ataxia should seek full genetic counselling. The risk of a woman with Friedreich's ataxia and no family history having an affected child is about one in 220 (pamphlet obtainable from the Friedreich's Ataxia Association, Burleigh Lodge, Knowle Lane, Cranleigh, Surrey).

1 Harding AE, Hewer RL. The heart disease of Friedreich's ataxia: a clinical and electrocardiographic study of 115 patients with an analysis of serial electrocardiographic changes in 30 cases. *Q J Med* 1983;208:489-502.

2 Hewer RL. Study of fatal cases of Friedreich's ataxia. *Br Med J* 1968;iii:649-52.

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Ingestion of button batteries: hazards and management

The widespread use of small, button sized batteries in household objects has led to increased awareness of the special hazards caused by their ingestion. A few reports have been published in the United States and Great Britain, but most have been isolated case reports, mainly of serious complications including death. Most authors have recommended early surgical intervention in all cases. We report our experience with three patients and review the previous reports.

Case reports

Case 1—A 6 year old boy swallowed the battery from a digital watch. Radiographs showed the intact battery in his stomach and, 24 hours later, in his caecum. He remained asymptomatic and passed the battery six days later.

Case 2—A 20 month old girl swallowed the battery from a calculator. Its presence was confirmed by radiography. She passed the battery intact three days later.

Case 3—A 4 year old girl swallowed the battery from a toy calculator. This was confirmed by radiography; repeat radiography one week later did not show any foreign body.

Comment

Before 1983 only six cases of ingestion of button batteries had been reported. Two of the patients died, one required a prolonged stay in hospital, two had suspected mercury poisoning, and in the remaining patient the battery perforated a Meckel's diverticulum.¹ Accordingly, many workers recommended early intervention.^{1,2} Litovitz, however, reviewed 56 cases in adults and children and recommended that surgical intervention should be withheld in the absence of specific clinical indicators.³

We reviewed the information available in reports on 58 children who ingested batteries. Forty eight were asymptomatic but, nevertheless, 16 underwent surgical intervention; seven of these showed evidence of minor mucosal damage. The remaining 32 asymptomatic patients were allowed to pass their batteries spontaneously. No late complications occurred in either group, which suggests that there is no benefit from surgical intervention in asymptomatic patients.

In five of the 10 patients with symptoms the battery lodged in the oesophagus; these five patients included the two who died and most of those with serious complications. The batteries ingested were 21-23 mm in diameter. These were the largest batteries in the entire series, and only one of the batteries that went beyond the oesophagus was of a similar size. The two patients who died had extensive liquefaction necrosis of the oesophagus, which led to severe mediastinitis in one and perforation of the aorta nine days after removal of the battery in the other. The three remaining patients suffered a second degree burn of the oesophagus (one) or tracheo-oesophageal fistula (one) or required a prolonged stay in hospital (one).

The battery went beyond the oesophagus in the five other patients with symptoms. Four of these batteries split, resulting in symptoms of mercury poisoning in two children. Two other patients with dark discoloration of

stools were managed conservatively and had no sequelae. Only one serious complication occurred in this group, when the battery lodged in a Meckel's diverticulum, causing liquefaction necrosis and perforation. This patient underwent laparotomy and survived.

On the basis of this information and our own experience we make the three following recommendations. Firstly, with few exceptions, ingestion of button batteries should be treated by observation alone. The obvious and important exception is when the battery is lodged in the oesophagus: this must always be treated as an emergency as oesophageal damage may occur within a few hours. Secondly, a chest radiograph should be obtained, especially if the battery is greater than 15 mm in diameter, as its size is the most important factor in determining whether it will pass beyond the oesophagus. Finally, if a battery that has gone beyond the oesophagus has split symptoms of mercury poisoning may occur and mercury concentrations should be measured.

1 Temple DM, McNeese MC. Hazards of battery ingestions. *Pediatrics* 1983;71:100-3.
2 Votteler TP, Nash JC, Rutledge JC. The hazards of ingested disc batteries in children. *JAMA* 1983;249:2504-6.
3 Litovitz TL. Button battery ingestions. *JAMA* 1983;249:2495-500.

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An unusual case of Gardnerella vaginalis septicaemia

Gardnerella vaginalis, first described in 1953 and originally thought to be a harmless commensal of the vaginal tract, has since been implicated as a cause of vaginitis,¹ bacteraemias,^{2,3} and various genitourinary infections in men.⁴ Many bacteraemias caused by *G vaginalis* in women, particularly post partum, have been reported. We report a case of *G vaginalis* septicaemia with severe endotoxic shock in a previously fit man.

Case report

A 19 year old man was admitted to hospital with a 24 hour history of fever, rigors, pain in the loins, dysuria, and general malaise. Two hours before admission he had become progressively more drowsy and unresponsive. He had been circumcised as a child and had since suffered occasional episodes of dysuria and frequency associated with a meatal stricture. He was feverish (41°C), had tachycardia (140 beats/min), and was hypotensive (90/60 mm Hg). There were no focal neurological signs, and septicaemia with shock secondary to a urinary tract infection was provisionally diagnosed. He was admitted to the intensive care unit, resuscitated with intravenous fluids, and treated with cefuroxime and gentamicin. Over the next 24 hours he developed disseminated intravascular coagulation, which was treated with platelets, cryoprecipitate, and fresh frozen plasma, and acute renal failure, which was treated with dopamine and mannitol. Over the next 10 days his renal function and clotting factors returned to normal, and he made a full and unremarkable recovery. Subsequent investigation of the renal tract showed a small bladder with thickened walls and diverticulae.

Blood samples and a midstream specimen of urine were taken on admission before antibiotic treatment and dilatation of his meatal stricture. Microscopy of the urine showed numerous pus cells and Gram positive bacilli, but no organisms grew from urine samples incubated for 24 hours on cystine-lactose electrolyte deficient medium. After incubation for 48 hours on blood agar anaerobic cultures of both blood and urine yielded a heavy growth of a small poorly α haemolytic colony. Gram staining showed the colonies to be variable but made up predominantly of Gram positive bacilli similar to those seen on urine microscopy. The organism did not produce oxidase or catalase but fermented glucose, maltose, and mannite and hydrolysed hippurate. It was resistant to a disc containing 2.5 µg metronidazole but was susceptible to a 50 µg disc. It was also sensitive to penicillin, ampicillin, and cephalosporins and resistant to tetracycline and cotrimoxazole. Despite the poor haemolysis these characteristics were consistent with those of *G vaginalis*. Indirect immunofluorescence of serum taken during the acute and convalescent phases showed an increase in antibody from a dilution of <1:8 to >1:128 against both the blood and urinary isolates.

Comment

There seems little doubt that this patient suffered a severe septicaemic illness caused by *G vaginalis* in his urinary tract. It has been estimated that *G vaginalis* is carried asymptomatically by 7-11% of men screened at a urology clinic.⁵ Several infections in men have implicated *G vaginalis*, including balanoposthitis, urethritis, cystitis, and asymptomatic bacteriuria.⁴ One case of bacteraemia also occurred after transurethral retropubic prostatectomy and was associated with fever, chills, and dysuria, and, though blood cultures were positive, urine culture was negative.³ The symptoms resembled those occurring in postpartum women rather than the severe endotoxic shock seen in our case. Carriage of the organism has been particularly associated with balanoposthitis, especially with a relative phimosis, and the meatal stricture seems to be the predisposing factor in this case. Although *G vaginalis* is usually carried asymptomatically in the male urinary tract, it is important to recognise that it may occasionally cause a severe systemic illness.

We thank the department of microbiology, Sheffield University Medical School, for its help in confirming the identity of the organism.

1 Pheiffer TA, Forsyth MS, Dunfee MA, et al. Non specific vaginitis. Role of Haemophilus vaginalis and treatment with metronidazole. *N Engl J Med* 1978;298:1429.
2 Reimer LG, Reller LB. Gardnerella vaginalis bacteraemia: a review of thirty cases. *Obstet Gynecol* 1984;64:170-2.
3 Patrick S, Garnett PA. Corynebacterium vaginale bacteraemia in a man. *Lancet* 1978;i:766.
4 Watson RA. Gardnerella vaginalis: genito urinary pathogen in men. *Urology* 1985;25:217-22.
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Red cell morphology at high altitude

In recent years the tourist industry has made areas at high altitude easily accessible. Exposure to high altitude may cause acute mountain sickness and its most severe complications—namely, high altitude pulmonary oedema and cerebral oedema.¹ The pathophysiology of acute mountain sickness is still unclear, but a contributing factor might be a change in the rheological properties of blood at high altitude. Recently, Rowles and Williams found an increased prevalence of abnormal red cell shapes, including echinocytes, in men who climbed above 4600 m for 39 days.² Such abnormal shapes, especially echinocytes, can cause an increase in blood viscosity.³ No data are available on the influence of short term exposure to high altitude on the morphology of red cells; this prompted our study.

Subjects, methods, and results

We studied 10 healthy volunteers (five women, mean (SD) age 30.4 (6.7) years, and five men, mean age 34.4 (5.2) years). Blood (100 µl) was drawn from an antecubital vein into a syringe containing 500 µl of 1% glutaraldehyde. The first blood sample was taken at an altitude of 1190 m before the subjects ascended to 3400 m in a cablecar. After an overnight stay at 3600 m they reached an altitude of 4560 m (Cabin Marguerita, Italy) by foot the next day. Blood was drawn at that altitude after a six hour rest in the cabin, 24 hours after the first sampling. The red cells were fixed for 48 hours and then examined by light microscopy at a magnification of ×800. They were classified blindly according to the classification of Bessis as discocyte; stomatocyte I (cup shaped (or convex-concave) red cell); stomatocyte II (red cell with a pronounced concavity); echinocyte I (irregularly contoured discocyte with up to five buckles); or echinocyte II (flat red cell with multiple spicules).⁴ The data were analysed with Student's *t* test for paired data.

The table shows the frequency distribution of the different types of cell. The discocyte is regarded as the equilibrium shape between two opposing transformations—that is, the stomatocytic and the echinocytic transformations.⁴ About 90% of the red cells were discocytes, and no difference was found in morphology at low and high altitudes.

Distribution of red cell shapes (%) in 10 subjects at 1190 m and, 24 hours later, at 4560 m according to classification of Bessis.⁴ Values are means (SD)

Altitude	Stomatocytes		Discocytes	Echinocytes	
	II	I		I	II
1190 m	1.0 (0.5)	11.9 (4.3)	86.9 (4.8)	0.3 (0.4)	0.1 (0.2)
4560 m	1.0 (0.6)	9.8 (2.6)	89.1 (2.4)	0.2 (0.2)	0